

GETTING THE FACTS

Hodgkin Lymphoma: Relapsed/Refractory

Hodgkin lymphoma (HL), also known as Hodgkin disease, is less common than non-Hodgkin lymphoma (NHL). Although HL can occur in both children and adults, it is most commonly diagnosed in young adults between the ages of 20 and 29 years. HL has been studied more than any other type of lymphoma, and because of the treatment advances over the past few decades, five-year survival rates have improved.

HL is often characterized by the presence of very large cells called Reed-Sternberg (RS). This type of lymphoma usually starts in the lymph nodes and can spread to other lymph nodes and, rarely, to other organs.

Common signs and symptoms of HL include swelling of the lymph nodes (which is often but not always painless), fever, night sweats, unexplained weight loss, itching, and lack of energy. While most people who have these complaints do not have HL, anyone with persistent symptoms should see a physician to make sure that lymphoma is not present.

COMMON TYPES OF HL

The two main classifications of Hodgkin lymphoma are classic HL (cHL, which accounts for over 90% of all cases) and nodular lymphocyte-predominant HL (around 5 percent of all cases).

The four subtypes of cHL are:

- Nodular sclerosis
- Mixed cellularity
- Lymphocyte-depleted
- Lymphocyte-rich

Treatment options for patients with nodular lymphocyte-predominant HL differ from those available to patients with cHL.

RELAPSED OR REFRACTORY DISEASE

For patients who *relapse* (disease returns after treatment) or become *refractory* (disease does not respond to treatment), secondary therapies are often successful in providing *remission* (disappearance of signs and symptoms) and may even cure the disease. For cHL, most relapses typically occur within the first three years following diagnosis, although some relapses occur much later. Patients who relapse often present with the same symptoms they had when first diagnosed with HL.

TREATMENT OPTIONS

A number of treatment options are available for patients with relapsed or refractory HL. The exact type of treatment prescribed for individual patients depends on several factors, including the timing of the relapse, age, overall health of the patient, scope of disease, and previous therapies received.

The standard treatment for patients with relapsed/refractory HL without other significant health conditions consists of *systemic* (throughout the body) therapy, usually followed by *autologous stem cell transplantation* (in which a patient's own stem cells are infused after high-dose chemotherapy). Involved-site radiation therapy (ISRT) may also be used. For more information on transplantation, view the *Understanding the Stem Cell Transplantation Process* publication on the Lymphoma Research Foundation's (LRF's) website (lymphoma.org/publications).

There are a variety of single-agent and combination therapy regimens that may be used for relapsed/refractory HL, including:

- Brentuximab vedotin (Adcetris)
- Bendamustine (Treanda)
- Nivolumab (Opdivo)
- Pembrolizumab (Keytruda)
- DHAP (dexamethasone, cisplatin, and cytarabine)
- ESHAP (etoposide, methylprednisolone, cisplatin, and cytarabine)
- GVD (gemcitabine, vinorelbine, and liposomal doxorubicin)
- ICE (ifosfamide, carboplatin, and etoposide)
- IGEV (ifosfamide, gemcitabine, and vinorelbine)

Brentuximab vedotin is approved for the treatment of relapsed/refractory cHL after failure of autologous hematopoietic stem cell transplantation (auto-HSCT), after failure of two previous chemotherapy regimens in patients who are not eligible for auto-HSCT, and as a *consolidation treatment* (a short and intense course of therapy to eliminate residual disease) after auto-HSCT in patients at high risk of disease relapse or progression. Nivolumab (Opdivo) is indicated for the treatment of adult patients with cHL that has relapsed or progressed after auto-HSCT and brentuximab vedotin or 3 or more lines of systemic therapy (including auto-HSCT). Pembrolizumab (Keytruda) is used for the treatment of adult and pediatric patients with relapsed or refractory cHL. In pediatric patients who have relapsed, pembrolizumab (Keytruda) is only approved after 2 or more lines of therapy.

TREATMENTS UNDER INVESTIGATION

In addition to conventional chemotherapies, there are several new agents currently being tested in clinical trials:

- AB-205 (E-CEL cells)
- Anti-CD30-chimeric antigen receptor (CAR) T cells
- Camidanlumab Tesirine (ADCT-301, Cami)
- Camrelizumab (SHR-1210)
- Everolimus (Afinitor)
- Ibrutinib (Imbruvica)
- Itacitinib (INCB039110)
- Ipilimumab (Yervoy)
- Lenalidomide (Revlimid)
- Magrolimab (5F9)
- Mocetinostat (MGCD103)
- Pralatrexate (Folotyn)
- Romidepsin (Istodax)
- Ruxolitinib (Jakafi)
- Tislelizumab (BGB-A317)
- Umbralisib (Ukoniq)

It is critical to remember that scientific research is continuously evolving. Treatment options may change as new treatments are discovered and current treatments are improved. Therefore, it is important that patients check with their physician or with LRF for any treatment updates that may have recently emerged.

CLINICAL TRIALS

Clinical trials are crucial in identifying effective drugs and determining optimal doses for patients with lymphoma. Because the optimal treatment of HL may vary for each patient, clinical trials are very important and will identify the best treatment options in this disease. Patients interested in participating in a clinical trial should view the *Understanding Clinical Trials* fact sheet on LRF's website (lymphoma.org/publications), talk to their physician, or contact the LRF Helpline for an individualized clinical trial search by calling (800) 500-9976 or emailing helpline@lymphoma.org.

FOLLOW-UP

Patients with lymphoma should have regular visits with a physician who is familiar with their medical history and the treatments they have received. Medical tests (such as blood tests and computed tomography [CT]/ positron emission tomography [PET] scans) may be required at various times during remission to evaluate the need for additional treatment.

Patients and their caregivers are encouraged to keep copies of all medical records and test results as well as information on the types, amounts, and duration of all treatments received. This documentation will be important for keeping track of any effects resulting from treatment or potential disease recurrences. LRF's award-winning *Focus on Lymphoma* mobile app (lymphoma.org/mobileapp) and *Lymphoma Care Plan* (lymphoma.org/publications) can help patients manage this information.

LRF'S HELPLINE AND LYMPHOMA SUPPORT NETWORK

A lymphoma diagnosis often triggers a range of feelings and concerns. In addition, cancer treatment can cause physical discomfort. The LRF Helpline staff members are available to answer your general questions about a lymphoma diagnosis and treatment information, as well as provide individual support and referrals to you and your loved ones. Callers may request the services of a language interpreter. A part of the Helpline is LRF's one-to-one peer support programs, Lymphoma Support Network. This program connects patients and caregivers with volunteers who have experience with HL, similar treatments, or challenges, for mutual emotional support and encouragement. Patients and loved ones may find this useful whether the patient is newly diagnosed, in treatment, or in remission.

MOBILE APP

Focus On Lymphoma is the first mobile application (app) that provides patients and caregivers comprehensive content based on their lymphoma subtype, including HL, and tools to help manage their lymphoma such as, keep track of medications and blood work, track symptoms, and document treatment side effects. The Focus On Lymphoma mobile app is available for download for iOS and Android devices in the Apple App Store and Google Play. For additional information on the mobile app, visit [FocusOnLymphoma.org](https://www.FocusOnLymphoma.org). To learn more about any of these resources, visit our website at [lymphoma.org](https://www.lymphoma.org), or contact the LRF Helpline at **800-500-9976** or helpline@lymphoma.org.

LYMPHOMA CARE PLAN

Keeping your information in one location can help you feel more organized and in control. This also makes it easier to find information pertaining to your care and saves valuable time. LRF offers a Lymphoma Care Plan as a resource for patients and their caregivers. LRF's Lymphoma Care Plan document organizes information on your health care team, treatment regimen, and follow-up care. You can also keep track of health screenings and any symptoms you experience to discuss with your health care provider during future appointments. The Lymphoma Care Plan document can be accessed by visiting [lymphoma.org/publications](https://www.lymphoma.org/publications).

Resources

LRF offers a wide range of free resources that address treatment options, the latest research advances, and ways to cope with all aspects of lymphoma and HL. LRF also provides many educational activities, including our in-person meetings, podcasts, and webinars for people with lymphoma. For more information about any of these resources, visit our websites at [lymphoma.org/HL](https://www.lymphoma.org/HL) or [lymphoma.org](https://www.lymphoma.org), or contact the LRF Helpline at **(800) 500-9976** or helpline@lymphoma.org.

Para información en Español, por favor visite [lymphoma.org/es](https://www.lymphoma.org/es). (For Information in Spanish please visit [lymphoma.org/es](https://www.lymphoma.org/es).)

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Supported through grants from:    

Understanding Lymphoma fact sheet series is published by the Lymphoma Research Foundation (LRF) for the purpose of informing and educating readers. Facts and statistics were obtained using published information, including data from the Surveillance, Epidemiology, and End Results (SEER) Program. Because each person's body and response to treatment is different, no individual should self-diagnose or embark upon any course of medical treatment without first consulting with his or her physician. The medical reviewer, the medical reviewer's institution, and LRF are not responsible for the medical care or treatment of any individual.

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Last updated 2022

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